The Initial Presentation of Distal RTA with Hypokalemic Quadriparesis in a male patient with Delayed Diagnosis of Primary Sjogren's Syndrome

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ABSTRACT

The exocrine glands are affected by Sjögren's syndrome, a systemic autoimmune illness characterized by lymphoplasma cell infiltration.[1] reduction of glandular activity over time. A secondary form that is linked to various autoimmune illnesses exists in addition to the primary form, which affects the exocrine glands with or without systemic involvement (scleroderma, rheumatoid arthritis, systemic lupus erythematosus, and connective tissue diseases, etc.) Renal Tubular Acidosis (RTA), which can be overt or latent and is caused by tubulo-interstitial nephropathy,it is the characteristic extra-glandular sign of primary Sjögren's syndrome.[2] Salivary or lacrimal gland biopsies are used to diagnose Primary Sjogren Syndrome

Key words-Distal Rta, Sjogren Syndrome, Hypokalemic Periodic Paralysis

INTRODUCTION

An exocrine condition affecting the salivary, parotid, lacrimal glands and other exocrine organs are called Sjögren's Syndrome, sometimes called Sicca Syndrome.[3] People with Sjögren's syndrome frequently experience interstitial nephritis and distal renal tubular acidosis (DRTA). Sjögren's syndrome affects non-exocrine organs like the kidneys.[4] However, Sjögren's syndrome patients seldom develop severe hypokalemia or paralysis brought on by Distal RTA. Despite the rarity of hypokalemic quadriparesis as the first sign of the condition, Sjögren's syndrome has previously been related to renal tubular acidosis and hypokalemia, Systemic Lupus Erythematosus, Rheumatoid arthritis, scleroderma are associated to a second variation that affects the exocrine glands and may or may not be systemic.[5]

CASE REPORT

A 31-year-old male presented with progressive weakness of limbs for 4 years, more in the upper than lower limbs, progressive and initially proximal. The patient was unable to walk on his own, lift his hands, or do household work. There was no history of dysphagia, diplopia, regurgitation of food, deviation of mouth, seizures, or loss of consciousness. The patient was completely bedridden for the last 2 months. For 4 years, the patient had intermittent improvements and worsening of the weakness. Initial Xrays PBH showed osteoporosis.USG Abdomen showed Right sided nonobstructive calculus in the kidney. MRI Whole spine revealed osteoporosis with mild posterior disc bulges without neurological compression. In two episodes S. Potassium Levels were found to be lower at 2.0 mmol/l and 1.8 mmol/l. 3 years ago patient once presented with quadriparesis and respiratory distress and a diagnosis of Myasthenia with Mysthenic Crisis was made. The patient was intubated and put on positive-pressure ventilation. At that point, the S. Potassium was 2.3 mmol/l.The patient recovered and was discharged

The patient again presented to Hospital, On presentation, the patient had weakness in both lower limbs and was bedridden. On examination, all Deep tendon reflexes were brisk. On Lab Investigations, Complete blood counts were normal except for ESR of 45 and S. potassium of 2.1. Renal and Hepatic functions were normal. Urinary Sodium was 50mmol/l and Urinary Potassium was 10mmol/l. S.Calcium was 9.6mmol/l. CRP was negative and CPK-Serum NAC levels were normal at 23. The thyroid profile was normal. Chest Xray Revealed generalized osteoporosis with looser's zones on the ribs. Nerve Conduction Studies were normal and EMG shows Neurogenic pattern myopathy. A diagnosis of Osteomalacia Myopathy was made and the patient was discharged.

The Patient again presented with similar complaints. On examination, The power was Grade 3/5 across all joints and waddling gait. There were no cerebellar signs. USG abdomen this time was suggestive of Medullary Nephrocalcinosis. Instead of normal renal functions, persistent hypokalemia, Nephrocalcinosis, and myopathy, Urinary pH and simultaneous ABG were done. It showed Normal Anion Gap Metabolic Acidosis with inappropriately high urinary pH.

Diagnosis of Distal Renal Tubular Acidosis was made. The patient was started on 30 ml of Scholl's Solution. The patient Drastically improved in one month and could walk on his own.

He admitted to experiencing dry eyes and mouth and was not able to chew food properly for two years.ANA by IF was Positive and ANA Blot showed the presence of SS-A and SS-B antibodies. Furthermore, USG of Parotid and Submandibular glands showed the possibility of Sjögren's syndrome, confirmed by labial Biopsy. Distal RTA was the clinical manifestation of Sjögren's disease, in this case, making it noteworthy. In individuals with RTA that appear to be idiopathic, we expect that Sjögren's syndrome will be taken into consideration.

USG

SALIVARY GLANDS- Bilateral parotid glands show multiple small well defined anechoic cysts with a reticular pattern, Bilateral submandibular glands appear atrophic, as seen in Sjogren Syndrome

Figure 1

Figure 2

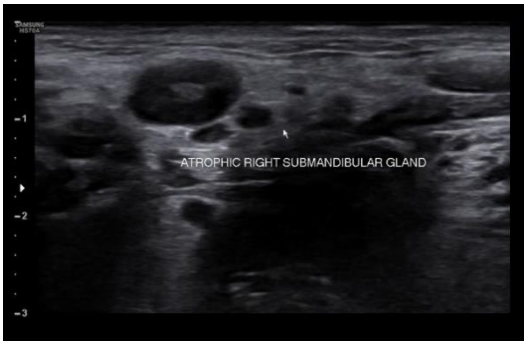


Figure 3

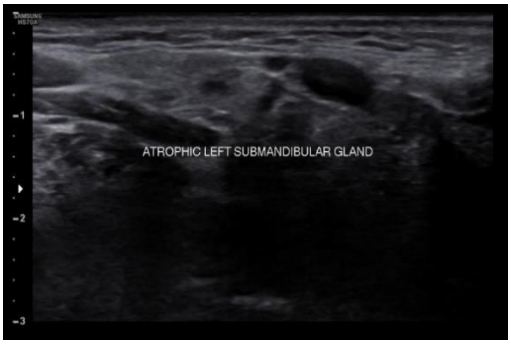
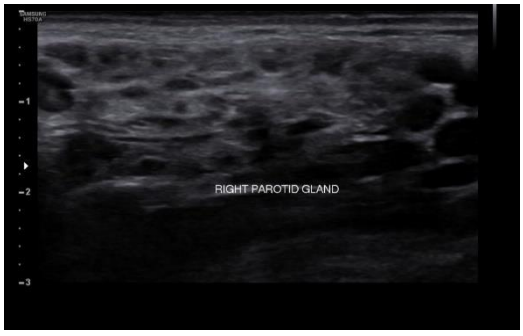
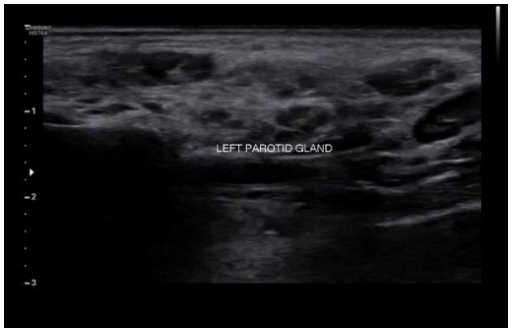


Figure 4





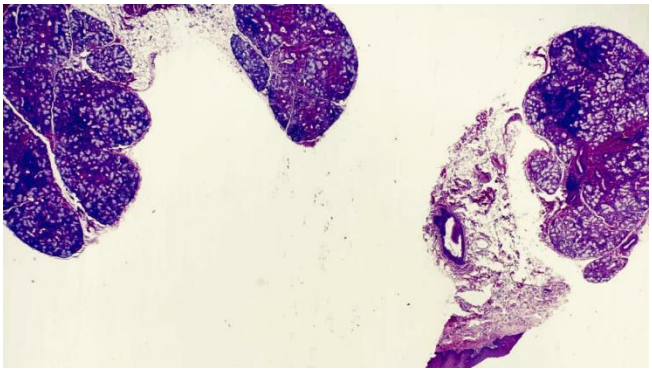


Figure 5

Numerous lobules of salivary gland tissue were seen in the lip biopsy (adequate; >4 mm2). There are six areas where more than fifty lymphocytes have been concentrated.

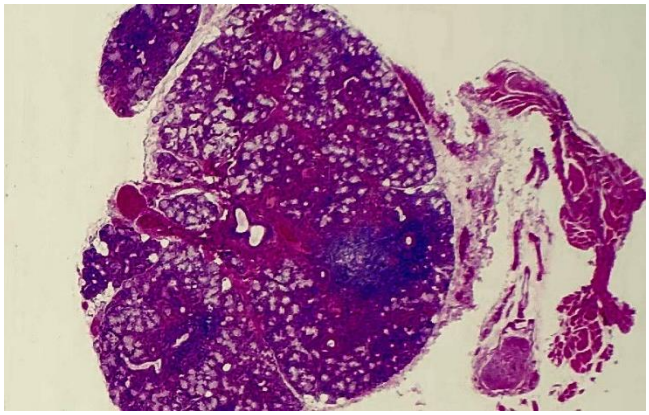


Figure 6

A large number of lymphocytes foci are seen in salivary gland tissue to support the diagnosis of Sjögren's syndrome

DISCUSSION

Distal RTA (dRTA) : In the presence of systemic metabolic acidosis , the kidneys cannot acidify the urine to a pH of 5.5 due to decreased hydrogen ion secretion or bicarbonate reabsorption in the distal nephron.[6] Normal anion gap metabolic acidosis, hypokalemia, bone demineralization, myopathy[7], respiratory distress, and even death are some of the consequences.

A multisystemic autoimmune illness called Sjogren's syndrome frequently manifests as lacrimal and salivary gland dysfunction. Although the earliest clinical sign of renal involvement in Primary Sjogren Syndrome is an uncommon occurrence, renal involvement has a prevalence of about 9[8] Tubulointerstitial involvement or, less frequently, glomerular involvement accounts for renal involvement. The pathophysiology of renal involvement has been attributed to plasma cell, T cell, or B cell lymphocytic infiltration of the renal tubules. Other causes include antibodies to the thiazide-sensitive NaCl cotransporter (NCCT) and

decreased hydrogen ion secretion as a result of the distal tubules' lack of a vacuolar H+ ATPase pump.[9]

CONCLUSION

As hypokalemia causes paralysis, the case under investigation might have been misled. However, the diagnosis of the illness was aided by supportive clinical signs and a positive antibody test, together with careful observation.

A proper set of guidelines and medication were used to handle the situation effectively. This type of clinical disease is not always easy to identify or diagnose with certainty. Since this disorder gradually worsens, it is important to promote awareness of its various presentational modalities. Such case studies can also be used to raise awareness by being distributed widely to all healthcare professionals.

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Conflict of Interest - None

Ethics approval and consent - All consents were taken from ethics committee

Patient consent for publication - Patient provided consent for his information to be published.

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